



Unusual presentation of Cardiac Myxoma

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General Note



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ABSTRACT

Cardiac myxomas are the most common type of benign primary cardiac tumours. Clinical presentations are usually due to haemodynamic, embolic or constitutional effects. We present a case of a 23 years old female, who presented to us with history of

weight loss, arthralgia and intermittent fever of 4 months duration. She was initially investigated for other causes of weight loss and fever. Her thoracic echocardiography revealed a left atrial mass of 6x5x4cm in left atrium, obstructing the mitral valve intermittently. After investigations patient underwent surgery under standard cardiopulmonary bypass for excision of tumour. Her postoperative course was uneventful. Patient gained 2kg of weight in next 2 months. The rarity of this case lies in atypical presentation of cardiac myxoma, where our patient presented with predominant constitutional symptoms rather than cardiac symptoms.

Keywords: Cardiac Myxoma, constitutional symptoms, thoracic echocardiography, mitral valve

1. INTRODUCTION

First report of Cardiac tumour appeared in 1559 (Mir et al., 2017). Cardiac myxomas are the most common benign cardiac tumours. Their clinical presentation depends on location, size, type and rate of growth. Sometimes there are bilateral cardiac tumours leading to more functional disturbances. Myxoma usually originates from multipotent mesenchymal stem cells or from neuroendocrine tissue. Sometimes myxomas can be malignant. Haemodynamic complications of cardiac myxoma are left heart failure and right heart failure. Embolic complications might be in systemic or pulmonary circulations. The most common clinical symptoms are dyspnoea, orthopnoea and paroxysmal nocturnal dyspnoea (PND). Clinical examinations in such cases usually reveal signs of left ventricular failure. The characteristic clinical sign which is found in auscultation is tumour plop which is heard during diastole. It is seen in around 50% of patients.

Constitutional symptoms of cardiac myxoma include malaise, anorexia, fatigue, fever, arthralgia and weight loss which mimics features of collagen vascular disease.

2. CASE REPORT

A 23 years old female presented to us with chief complaints of 8kg weight loss over a period of 4 months, intermittent myalgias, arthralgias and fever since 4 months, syncopal attack twice in 15 days. There was no history of cough, expectoration, dyspnoea, orthopnoea, PND, hemoptysis, swelling in joints, morning stiffness, back pain, night sweats and no history of seizure.



Figure 1 transthoracic echocardiography showing large left atrial mass.

On examination patient was found to be of thin built, conscious, comfortable in sitting position, pulse was 88/min, regular and all peripheral pulsations were felt. Blood pressure - 120/70 mm of Hg. Pallor was present. There was no icterus, cyanosis, clubbing or lymphadenopathy. JVP was normal. Cardiovascular system examination was normal. CNS, RS and per abdomen examination was also normal.

Investigations revealed complete blood count – Hb 8gm/dl, normocytic normochromic RBCs on PS, TLC 8700/mm³ with 66% neutrophils. ESR – 70mm in first hour, Platelet count – 6.5 lacs, C-reactive proteins – 15mg/dL, thyroid profile was normal, serum ANA, dsDNA, C-ANCA were negative, RA factor was negative, serum Interleukin-6(IL-6) level was 28pg/ml (normal 5-15pg/ml). Mantoux test was non-reactive. X-ray chest – lung fields were normal but showed straightening of left atrial border. Due to history of syncope, CT of brain was done which was normal. A transthoracic echocardiography was done which showed large left atrial mass 6x5x4cm, popping in and out of mitral valve, with severe mitral stenosis with mitral regurgitations with moderate pulmonary hypertension and mild tricuspid regurgitation (fig 1). Ejection fraction was 50%. No other mass was seen anywhere in heart. Patient was not ready for cardiac CT. After stabilizing, patient was taken for open heart surgery.



Figure 2 Histopathological gross specimen showing 6x5x4cm of irregular tissue piece with soft gelatinous outer surface.

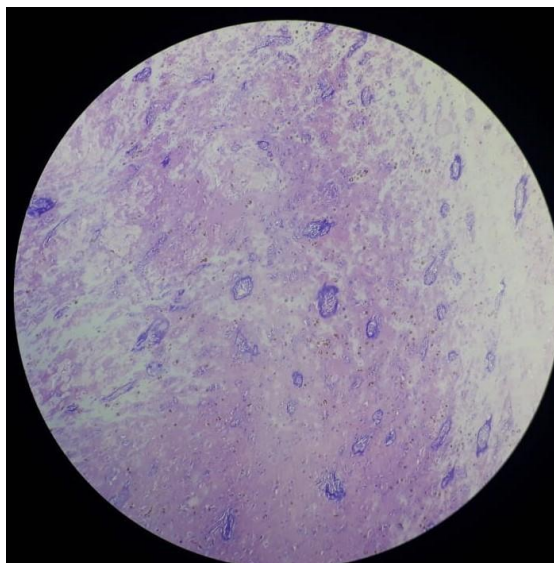


Figure 3 Microscopic section showing nests and cords of myxoma cells embedded in abundant amount of myxoid material. Myxoma cells are round to oval, having abundant amount of cytoplasm, nucleus is oval in shape and size with no nucleoli.

During surgery invasive monitoring was done with internal jugular cannulation, radial and femoral cannulation for Invasive Blood Pressure (IBP) and Trans esophageal Echocardiography (TEE). Induction of patient was done in Trendelenburg's position. Excision of the left atrial pedunculated mass was done under standard Cardiopulmonary Bypass. Atrial Septal Defect (ASD) was closed with pericardial patch. After closure of the heart during TEE Exam mitral valve regurgitation was noted so patient was opened again and mitral valve repair was done.

Histopathological gross specimen showed, 6x5x4cm of irregular tissue piece and soft gelatinous outer surface (fig 2). On cut section the surface was greenish gelatinous in appearance along with brown colour haemorrhagic areas. Histopathological and Microscopic findings confirmed atrial myxoma (fig 3). Her postoperative course was uneventful and was discharged without any anticoagulation.

3. DISCUSSION

Cardiac myxomas are non-cancerous primary tumours of heart. They constitute about 50% of all primary heart tumours (Thyagrajan et al., 2017). They are inherited as autosomal dominant character (Boutayeb et al., 2017). They are mostly intracavitary tumours. 75-80% in left atrium, 15-20% in right atrium, 34% in right or left ventricle and are common in adult females. Myxomas may be solitary or multiple. Size of the myxomas varies from 1-15cm; they may be globular or papillary with stalk. Papillary are more friable with more incidence of embolization. They are soft, gelatinous in consistency with areas of necrosis and haemorrhage.

Constitutional symptoms of myxomas are fatigue, fever, weight loss. Elevated C-reactive proteins, elevated IL-6 and elevated plasma globulins are present in these patients. Our patient had weight loss and increased C-reactive proteins. IL-6 is secreted by tumour which increases C-reactive proteins and gamma globulins (Yuan et al., 2017). Endo et al., 2002; in their study of constitutional symptoms and myxomas observed that 31% patient had fever and weight loss, 39% patient had increased CRP level and 21% patient had hypergammaglobulinemia. At least one constitutional symptom was present in 49% of the patient (Endo et al., 2002). All constitutional signs disappeared after myxoma surgery but not with malignant tumours. Large tumour size and multicentric origin was also observed in these patients. IL6 was not increased in patients without constitutional symptoms. IL-6 can affect septal and inferior wall of left ventricle leading to hypokinesia and heart failure (Dixit et al., 2017). Hypokinesia improves after a section. Our patient also had increased IL-6 level.

Transthoracic echocardiography can easily diagnose location, size of myxoma. CT Pulmonary Angiography (CTPA) and cardiac MRI can differentiate cardiac masses and pulmonary emboli. Differential diagnosis will be according to presenting symptoms. Our patient was investigated for tuberculosis, malignancies, thyrotoxicosis, collagen disorders, and collagen vascular disease. During surgery patient should be induced in Trendelenburg's position, the vent should be inserted after aortic clamping in case of left atrial myxoma (Mandryk et al., 2018). Median sternotomy is the standard surgical approach. Other technique is by right anterolateral mini-thoracotomy which reduces the hospital stay and favourable cosmetic outcome. Cryoablation at tumour site was also used to prevent recurrence. Normal recurrence rate is 1-3% while with familial predisposition risk of recurrence is upto 25% (Boutayeb et al., 2017). Cardiac manipulations should be minimal to avoid embolic complications.

4. CONCLUSION

It is rare to have cardiac myxomas only predominantly presenting as constitutional symptoms. Diagnosis can be delayed in such cases. In our patient syncope was the only cardiac manifestation of myxoma. Any patient presenting with features of weight loss, fever, arthralgia should undergo echocardiographic evaluation to rule out occult cardiac myxomas. Surgical excision is the best option available for treatment as symptoms disappear after surgical resection and long term prognosis is good. The need of long term anticoagulation is controversial. Follow up with cardiac echocardiography is must.

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